Aortic Intramural Hematomas and Penetrating Aortic Ulcerations:

Indications for Treatment Versus Surveillance

A review of the currently available literature focused on categorizing intramural hematomas and penetrating aortic ulcerations and when to treat them.

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Acute aortic syndrome (AAS) encompasses an array of potentially life-threatening aortic pathologies, including dissection, intramural hematoma (IMH), and penetrating aortic ulcer (PAU). AAS includes a heterogeneous group of patients; however, the underlying pathology universally involves disruption of the aortic intima and media. These diagnoses frequently overlap, and the initiating event may be difficult to pinpoint when more than one of these processes are seen together. Characteristics associated with elevated risk of PAU or IMH include male sex, hypertension, and presentation with chest pain. AAS represents potentially life-threatening disease processes, and therefore expeditious diagnosis, anatomic characterization, and treatment with hemodynamic control and/or intervention are essential in minimizing the morbidity and mortality associated with these conditions.

PAUs and IMHs involve the thoracic aorta in approximately 60% to 70% of cases, with PAUs frequently diagnosed incidentally outside of the acute phase.[1] PAU can evolve into an IMH, dissection, pseudoaneurysm, or aortic rupture. One in eight patients with acute aortic dissection may have a component of PAU or IMH.[2] The risk of aortic rupture has been demonstrated to be higher (40%) with an aortic dissection with PAU than with type A dissection alone (7%) or type B dissection alone (3.6%).[3] The rupture rate of PAU alone without aortic dissection ranges from 4.1% to 38%, and it can be as high as 18% to 26% for IMH alone without aortic dissection in acute presentations.[1,4-6] Initial management of PAU and IMH involves close hemodynamic monitoring, with medical treatment aiming to control blood pressure (BP) and heart rate. Close monitoring for changes in signs and symptoms as well as interval reimaging are essential in guiding appropriate decision-making on the need for intervention.

PENETRATING AORTIC ULCER

First diagnosed as a unique condition in 1986 by Stanson et al, an isolated PAU is characterized by disruption of the arterial intima and elastic lamina that extends into the media within the atherosclerotic aorta.[7] This disruption results in a focal outpouching through an area of vascular calcification with associated arterial flow and is most often noted on CTA (Figure 1).[1,2] PAU is frequently diagnosed incidentally, outside of the acute phase. An isolated PAU may have an indolent clinical course, as in the setting of the asymptomatic, stable, incidentally noted ulceration, or it may enlarge or develop associated IMH, dissection, pseudoaneurysm, or rupture.[1-7] Some PAU cases may present with associated IMH, as characterized by the presence of an ulcer-like projection in conjunction with surrounding hematoma extending along the aortic wall seen on CT. Isolated PAUs are seen in 2.3% to 7.6% of AAS cases and can be identified in all segments of the aorta; however, they are more common in the thoracic than the abdominal aorta and are most common in the descending

thoracic aorta (62%).[2,8] PAU may occur in a solitary location or in multiple segments of the aorta; however, when the ascending aorta is involved, rupture or concomitant IMH are more common.[2,9] In a single-institution review of PAU, the incidence of rupture on presentation was 4.1%, and endovascular or open repair was required in 12.9%.[1] The rupture rate has been reported to be as high as 38% for PAU in an acute presentation, which is considerably higher than that seen for aortic dissection.[4] Indications for repair include persistent or recurrent symptoms, rupture, or development of pseudoaneurysm. The indications for intervention versus observation for asymptomatic PAU is still unclear, with some authors advocating against repair in the acute setting.

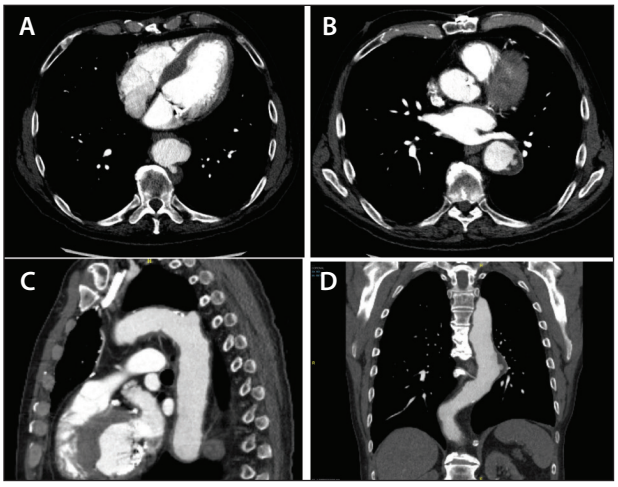
INTRAMURAL HEMATOMA

IMH is defined as disruption in the intima of the aortic wall leading to penetration of the media and accumulation of blood within the wall, without the identifiable flap associated with true aortic dissection.[10-12] The location of the intimal disruption may be difficult to visualize given that intramural blood is thrombosed (Figure 2). This characteristic differs from the persistent false lumen flow characteristic of aortic dissection. IMH can coexist with PAU in 45% of cases and may develop into progressive aortic dissection or aneurysm.[11,12] The incidence of IMH has been documented in 5% to 20% of all AAS cases.[2,12] IMH may resolve in nearly all patients who undergo endovascular intervention. Progression to aortic dissection has been reported in 28% to 74% of patients with IMH, and up to 20% to 45% of patients develop aneurysmal degeneration with or without a contained rupture.[13]

CLASSIFICATION

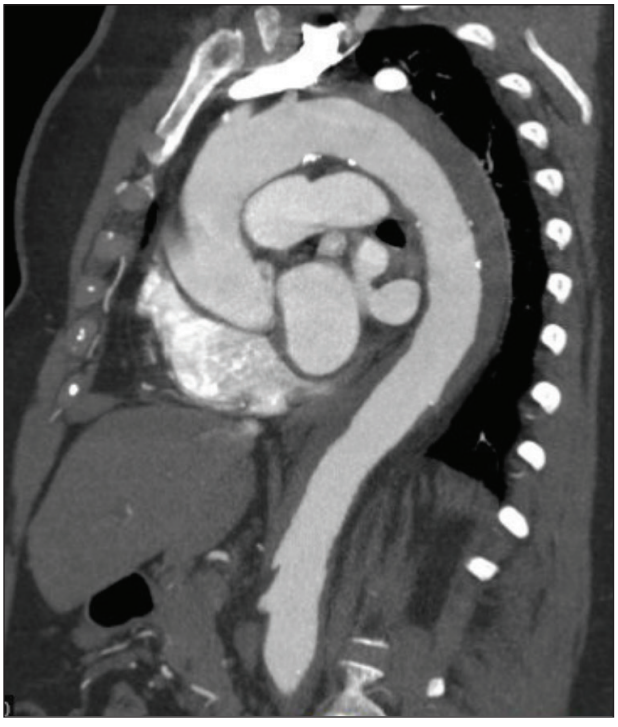
DeBakey and Stanford classifications have historically been the most commonly used aortic dissection classification systems. Stanford classification takes into consideration the presence (type A) or absence (type B) of ascending aortic involvement. DeBakey classification is based on the site of origin and termination of the dissection, with type I originating in the ascending aorta and extending past the aortic arch, type II originating and terminating in the ascending aorta, and type III originating in the descending aorta and extending distally.[14,15]

Similarly, PAU and IMH can be classified into presence (type A) or absence (type B) of ascending aortic involvement.[12] Type A IMH involves the ascending thoracic aorta or aortic arch, and type B IMH involves the descending thoracic aorta distal to the takeoff of the left subclavian artery. Due to the high mortality rate (55%) when managed medically, a type A IMH is typically repaired with emergent surgical intervention.[12,16] Type B PAU and IMH may be managed more conservatively than aortic dissection, with overall similar outcomes for dissection in the same anatomic distribution.[12] Recently, new reporting standards have been described for type B aortic dissections that take into account the chronicity of the dissection in addition to the presence of concomitant aortic processes such as IMH and PAU.[17]



Courtesy of Carlos Bechara, MD.

Figure 1. PAU of the descending thoracic aorta in axial (A, B), sagittal (C), and coronal (D) views.



Courtesy of Paul Crisostomo, MD.

Figure 2. IMH of the descending thoracic aorta.

DIAGNOSIS

Prompt diagnosis of AAS is imperative given the 30-day mortality rate of > 50% for aortic dissection and its associated findings.[18,19] Diagnosis of AAS is best made with the use of CTA of the chest and abdomen, ideally including the region extending from 3 cm above the aortic arch to the femoral vessels bilaterally. MRI may be more sensitive in detecting small PAU or IMH; however, contrast enhancement (ie, MRA) is essential for detecting small intimal defects.[2] Transesophageal echocardiography may also be used to assist in the diagnosis of ascending aortic and arch pathology.[20]

Common clinical presentations include chest pain, back pain, and hypertension.[1,2] Pain is the most common clinical presentation and results from stimulation of the aortic nerve plexus by rapid expansion.[11,21] AAS-related pain is typically described as severe, intense, acute-onset searing or tearing and is sometimes throbbing in nature. Pain in the distribution of the anterior chest, neck, and jaw may indicate ascending aortic involvement, whereas back and abdominal pain more frequently indicate thoracic involvement.[21] Although rare, distal embolization may be a presenting finding of PAU if thrombus forms on its surface, and it is more common in abdominal aortic than thoracic PAU.[22] Reported characteristics of the pain may change as the pathology of PAU and IMH evolves. Risk factors for the development of AAS include age ≥ 70 years, male sex, hypertension, cocaine or other stimulant use, atherosclerosis, previous aortic operation, previous catheter-based interventions, bicuspid aortic valve, and connective tissue disorders.[1,2,12] One study documented that approximately 9% of patients were asymptomatic on presentation.[11]

When IMH or PAU are diagnosed, initial workup should include an electrocardiogram, chest x-ray, and basic blood work (creatinine, complete blood count, liver function tests, toxicology screen, type and screen, D-dimer, and troponin) to rule out concomitant acute coronary syndrome. Chest x-ray may demonstrate a pleural effusion or widened mediastinum or may appear normal, but these findings would also be seen on CTA.[18]

Predictors of complications in the setting of AAS are recurrent symptoms, persistent pain despite adequate BP control, presence of PAU, involvement of the ascending aorta, end-organ ischemia, aortic diameter ≥ 5 cm, enlarging aortic diameter during surveillance interval, aortic wall thickness > 11 mm, recurrent pleural effusions, and difficult BP control.[1,13,16,23] An IMH in patients with an associated PAU has been demonstrated to progress more frequently than an IMH without PAU (48% vs 8%).[12]

MEDICAL MANAGEMENT AND INDICATIONS FOR INTERVENTION

Initial treatment of PAU or IMH should aim to decrease aortic wall stress to reduce the likelihood of progression to dissection or rupture. Pain control is an essential component of initial and ongoing management because analgesics can aid in decreasing the sympathomimetic-related increase in heart rate seen in these patients.[24] In symptomatic patients with PAU or IMH, intensive care unit admission is warranted for close hemodynamic monitoring. Tight control of heart rate and BP with goals of 60 to 80 bpm and 100 to 120 mm Hg systolic, respectively, are the mainstays of hemodynamic control.[2,24,25] The first line of treatment includes a tailored approach with intravenous β-blockers (propranolol, metoprolol, labetalol, esmolol). Non-dihydropyridine calcium channel blockers such as verapamil or diltiazem may be considered in patients who are intolerant to β-blockers. Vasodilators (eg, sodium nitroprusside) are second-line agents that may be added to β-blocker regimens to achieve rapid BP optimization.

Thoracic endovascular aortic repair (TEVAR) is recommended for complicated AAS, including dissection defined by persistent or recurrent pain, uncontrolled hypertension despite aggressive medical therapy, aortic expansion, malperfusion, or rupture. The presence of hypertension and symptoms on presentation predicts eventual need for repair. In a study by Nathan et al, 36% of such patients required repair, even if the PAU was initially treated with conservative management resulting in resolution of pain.[1] Indications for repair include pseudoaneurysm with diameter > 2 cm, rupture, dissection, hemodynamic instability, organ ischemia, failure to adequately respond to antihypertensive treatment, maximum aortic diameter > 55 mm, PAU base > 20 mm or depth > 15 mm, IMH with significant periaortic hemorrhage, or persistent or recurrent symptoms.[1,20,26,27] The method of repair can be TEVAR with carotid-subclavian bypass, endovascular aneurysm repair, or open surgery if there is involvement of the aortic arch and visceral vessels.[1]

Significant predictors of PAU progression as seen on serial imaging are symptomatic status and PAU depth > 15 mm, which has been found to be an independent predictor of mortality.[1,27] In the cohort evaluated by Nathan et al, 43% of symptomatic patients had radiographic progression in the surveillance interval, highlighting the importance of close surveillance in this population.[1] Of note, there was no difference in disease progression between PAUs, PAUs with saccular aneurysms, or PAUs with IMH.[1] A PAU with a 10-mm neck has also been suggested to be an indication to undergo early surgical intervention.[13] There was no difference in disease

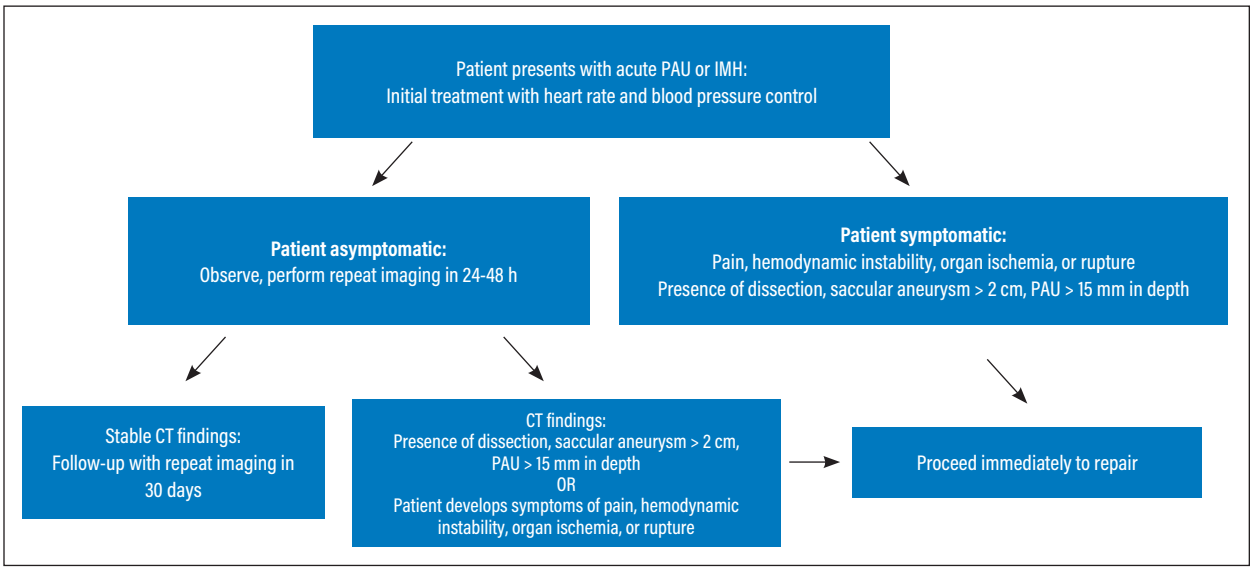


Figure 3. Flow diagram demonstrating steps in management of patients diagnosed with PAU or IMH.

progression between type B PAUs, PAUs with saccular aneurysms, or PAUs with IMH in the study by Nathan et al; however, < 20% of PAUs were symptomatic on presentation.[1] A flow diagram to help manage patients with symptomatic and asymptomatic PAU and/or IMH is shown in Figure 3.

Due to the high associated mortality rate, PAU and IMH involving the ascending thoracic aorta warrant emergent surgical intervention.[16] Traditionally treated with open surgical repair, endovascular repair may be possible in as many as 20% to 30% of cases, but these patients must be deemed inoperable for open repair and require treatment at a highly specialized center.[2] As there are currently no commercially available endovascular devices for treatment of the ascending aorta, strategies for repair include proximal aortic cuffs, snorkeling parallel grafts, and surgeon-modified devices. Uncomplicated ascending aortic hematomas may be managed medically, but as many as 45% of these patients may progress to aortic enlargement or dissection, so close observation is imperative.[28]

Treatment of PAU and IMH in the descending thoracic aorta usually includes endovascular stent grafting when possible, with carotid-subclavian bypass if coverage of the left subclavian artery is necessary because open operative repair has a mortality rate of 16%.[1,6,13,24] Longer stent grafts (20-25 cm) are often preferred to cover all areas of intimal disruption.[26] Figure 4 shows aortic remodeling that is common after treatment of an IMH with a TEVAR device at 1-month follow-up.

A recent literature review on TEVAR of AAS over the last 10 years indicates a 30-day mortality rate of only 4.8%, technical success rate of 98.3%, and early morbidity rate of 36.4%, the majority of which were access or endoleak related.[11,27] The left subclavian artery was covered in 21.8% of cases, and spinal drains were placed preventively in 24.7% of cases, with neurologic events of stroke and paraplegia occurring in only 2.4% of patients.[11] Eighty percent of cases used only one TEVAR device, and mean in-hospital length of stay was 7 days.[11] Survival rates after TEVAR for PAU were 91.1%, 79.3%, and 67.3% at 1, 3, and 5 years, respectively.[11] This led the authors to conclude that patients with asymptomatic incidental findings of PAU should be observed, whereas patients with recurrent or persistent symptoms despite optimal medical therapy, aortic diameter > 5.5 cm or an increase of ≥ 10 mm in size growth per year, and development of dissection, aneurysm, rupture, fistula, or IMH should undergo TEVAR repair.[11]

Abdominal aortic PAUs that are diagnosed incidentally can often be safely observed. Flohr et al performed a retrospective analysis of incidentally found PAU of the abdominal aorta and concluded that long-term mortality of this population was high; however, there were no observed ruptures in their 36-month follow-up data, and there was no difference in mortality or aortic pathology in patients with rates of growth > 1 mm per year versus < 1 mm per year.[8]

For patients with incidentally found aortic PAU or IMH or those who become asymptomatic with medical management alone, close follow-up is imperative. After hospital discharge on an appropriate medical regimen for heart rate and BP control, repeat CTA at 1, 3, 6, and 12 months should be performed, with annual surveillance thereafter if stable.[24] Patients who have undergone

Courtesy of Paul Crisostomo, MD.

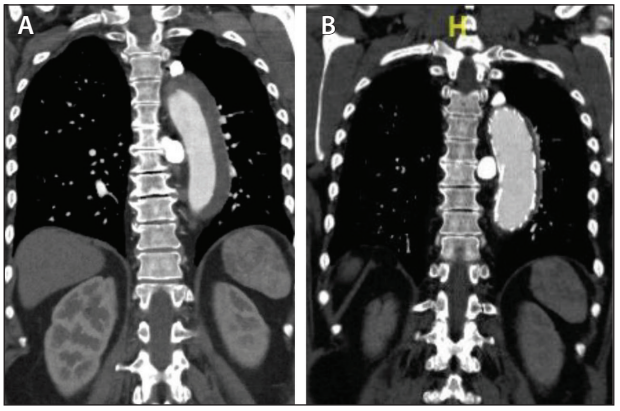


Figure 4. IMH of the descending thoracic aorta preintervention (A) and after endograft repair with aortic remodeling (B).

endovascular repair with a stent graft or open repair for isolated PAU or IMH should also undergo continued surveillance with CTA.

CONCLUSION

PAUs and IMHs are often seen together or in conjunction with acute aortic dissection. When diagnosed in the symptomatic patient, these complex aortic pathologies represent a potentially life-threatening medical condition. Prompt identification, medical management, and patient selection for intervention are critical components of care, along with long-term surveillance. ■

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